

Ventricular Inversion With Normal Ventriculoarterial Connection and Left Atrial Isomerism: Correction by the Mustard Operation

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A 2 year old boy with ventricular inversion and normal ventriculoarterial connection is described. Associated cardiac lesions included single atrium, absence of the coronary sinus, bilateral superior venae cavae, absence of the hepatic segment of the inferior vena cava with azygos and hemiazygos continuation, right aortic arch, levocardia and left atrial isomerism. At 5 days of age, the patient underwent a Waterston (aortopulmonary)

anastomosis because of suspected pulmonary atresia. The correct diagnosis was established at 2 years of age and the patient had a successful Mustard operation (interatrial baffle procedure) and closure of the Waterston anastomosis. Accurate preoperative diagnosis is difficult in this rare cardiac anomaly and the reported mortality rate is high.

The term ventricular inversion has been used to describe a discordant connection between the atria and ventricles. This anomaly is usually associated with ventriculoarterial discordance (corrected transposition of the great arteries), but normal ventriculoarterial connection has been reported in 14 cases (1-10). This complex has been termed isolated ventricular inversion (3,7), ventricular inversion without transposition (5,11) and clinical complete transposition of the great arteries (12).

The purpose of this report is to note the previously undescribed association of left atrial isomerism with this anomaly and review the anatomic, clinical and laboratory findings and surgical treatment.

Case Report

This Caucasian boy was first referred to Children's Hospital of Pittsburgh at 2 years of age. He had been markedly cyanotic shortly after birth and underwent cardiac catheterization elsewhere at 3 days of age. He was thought to have pulmonary atresia, and a Waterston (aortopulmonary) anastomosis was performed at 5 days of age. Postoperatively

cyanosis lessened, but growth and development were delayed. The patient was receiving digoxin and furosemide.

Clinical features. On admission, weight was 10.9 kg (5th percentile) and height was 85.5 cm (10th percentile). There was minimal cyanosis of the lips and nailbeds, but no clubbing. There was a prominent thrill in the suprasternal notch and at the high left sternal border. The second heart sound was loud and single. There was a grade 4/6 harsh pansystolic murmur at the high left sternal border and a grade 2/6 medium-pitched mid-diastolic murmur at the low left sternal border. The liver was midline. Peripheral pulses were prominent, but not bounding. Hemoglobin was 10.5 g/100 ml and hematocrit was 37%.

The electrocardiogram demonstrated sinus rhythm with an upright P wave in lead II. The QRS axis was 90°. A Q wave in the right precordial lead and absent Q waves in leads V₅ and V₆ suggested ventricular inversion. Evidence of right ventricular hypertrophy was seen.

Chest roentgenography demonstrated the cardiac apex to the left and the stomach gas bubble on the right. There were a right aortic arch, generalized cardiomegaly and increased pulmonary vascular markings. An overpenetrated chest roentgenogram showed bilateral left bronchi.

Two-dimensional echocardiographic study demonstrated two distinct atrioventricular (AV) valves and two well formed ventricles (Fig. 1A and B). The ventricular septum was intact, but the atrial septum could not be identified. The great arteries were normally related; the pulmonary artery was located anterior to the aorta (Fig. 1C and D). The aorta

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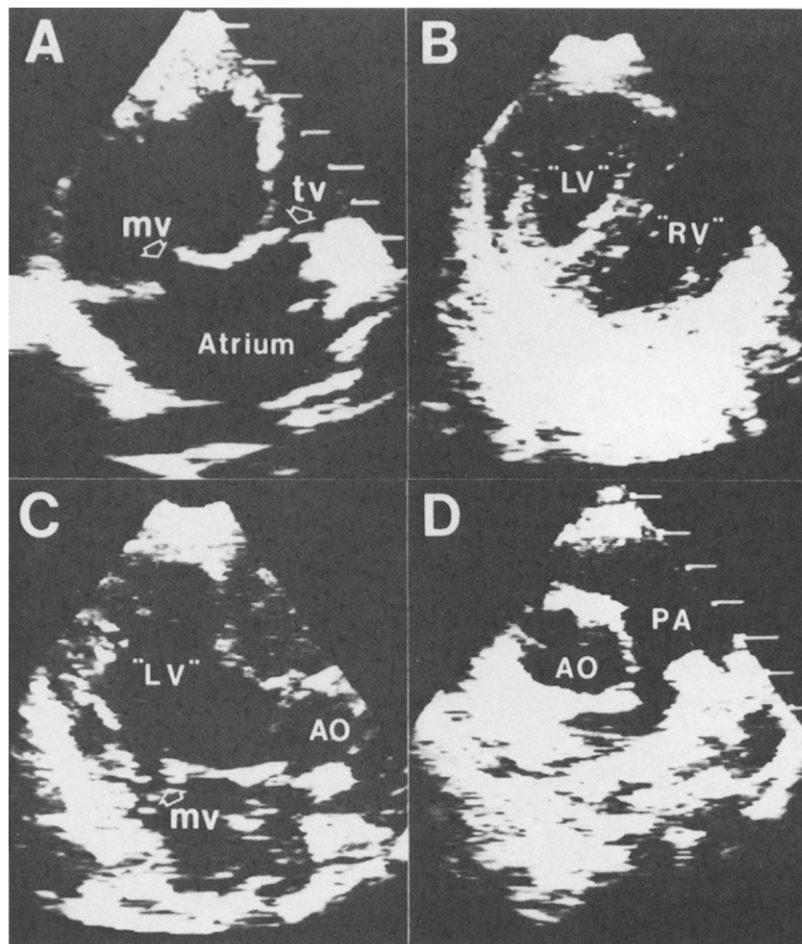


Figure 1. Two-dimensional echocardiograms. **A**, Four chamber view: the left-sided AV valve is more apically placed than the right-sided valve, suggesting that it is the tricuspid valve (tv) and that the right-sided valve is the mitral valve (mv). **B**, Short-axis view at the midventricular level: a circular, right-sided morphologic left ventricle ("LV") and crescent-shaped, left-sided morphologic right ventricle ("RV") are seen. **C**, Anteriorly angled projection from the four chamber view. Aortic-mitral valve (mv) continuity is seen. **D**, Short-axis view at the great artery level. A posteriorly located aorta (AO) and anteriorly located pulmonary artery (PA) with normal bifurcation are seen.

originated from a right-sided, apparently morphologic left ventricle with aortic-mitral valve continuity.

Catheterization findings. Cardiac catheterization was performed and hemodynamic data indicated moderate pulmonary hypertension and a pulmonary to systemic flow ratio of 2.5:1 (Table 1). On cineangiography, the right-sided anterior ventricle was morphologically a left ventricle from which the aorta arose (Fig. 2A and B). Aortic-mitral valve continuity was present. The left-sided posterior ventricle was morphologically a right ventricle (Fig. 2C and D). The outflow tract of this ventricle coursed anteriorly around the right-sided ventricle and gave rise to the anterior pulmonary artery. The aortic arch was right-sided and the descending aorta was located on the right side of the spine. There was considerable flow through the aorticopulmonary shunt. The inferior vena cava was to the left of the spine below the diaphragm and then divided into two branches; the larger branch drained into a left superior vena cava and then into the left side of the atrium while the smaller branch drained into a right-sided superior vena cava to the right side of the atrium. Both atrial appendages had left atrial morphology. Hepatic veins drained into the right side of the atrium.

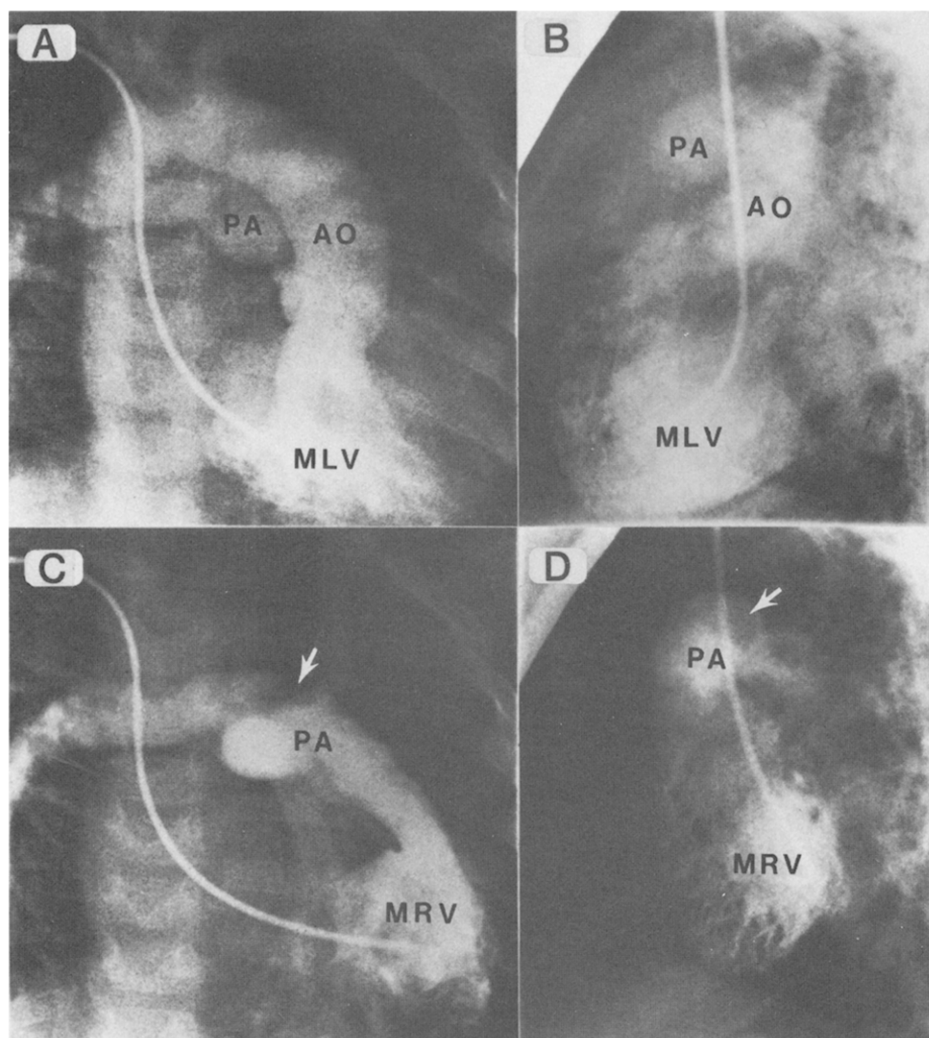
Surgery. The patient underwent a Mustard operation, and closure of the Waterston shunt using deep hypothermia and circulatory arrest. External inspection of the heart showed two left atrial appendages (Fig. 3). The anterior and dilated main pulmonary artery arose from the left-sided ventricle. The left pulmonary artery originated from the posterior aspect of the main pulmonary artery and coursed behind the

Table 1. Hemodynamic Data at Cardiac Catheterization

Site	Pressure (mm Hg)	Oxygen Saturation (%)
SVC	—	62
Atrium	m = 16	86
MLV	100/0	89
Aorta	85/45 (m = 60)	86
MRV	65/0	88
LPA	55/35 (m = 45)	—
RPA	60/40 (m = 46)	86
RPA wedge	m = 16	96

LPA = left pulmonary artery; m = mean; MLV = morphologic left ventricle; MRV = morphologic right ventricle; RPA = right pulmonary artery; SVC = superior vena cava (right-sided).

Figure 2. Angiocardiography with selective injection into the right-sided ventricle (A, anteroposterior; B, lateral) demonstrates a smooth-walled morphologic left ventricle (MLV) supporting the aorta (AO). The pulmonary artery (PA) is anterior to the aorta and fills via the Waterston shunt. Selective injection into the left-sided ventricle (C, anteroposterior; D, lateral) demonstrates a heavily trabeculated morphologic right ventricle (MRV). The pulmonary artery fills from this ventricle. The radiolucent area in the main pulmonary artery (white arrows) is due to washout from the Waterston shunt.



ascending aorta. Inspection of the intracardiac anatomy through a transverse right atrial incision showed no interatrial septum or coronary sinus. There were two separate pulmonary veins from the right lung, but only a single confluent pulmonary vein from the left lung. The left-sided posterior AV valve was anatomically a tricuspid valve and the right-sided anterior AV valve had the appearance of a mitral valve. Both AV valves were of normal size, without abnormality. An interatrial baffle procedure (Mustard operation) was performed using glutaraldehyde-preserved bovine pericardium.

The patient had an uneventful postoperative course and was discharged 10 days after operation. At follow-up 2 months later, the patient was well and without symptoms.

Discussion

Only 14 cases of ventricular inversion with ventriculo-arterial concordance have been reported (10); the diagnosis

was confirmed at autopsy in 13 and at cardiac catheterization and open heart surgery in 1. The female/male sex ratio was 2:1. All the patients manifested cyanosis at birth, and five had congestive heart failure that required medical treatment. All patients had cardiomegaly with increased pulmonary vascular markings except for two with proven pulmonary stenosis. Electrocardiograms were described in 10 patients; the findings were variable and of no value in diagnosis.

The echocardiographic findings of this anomaly have not been described previously. In our case, two-dimensional echocardiography was diagnostic and unique, at least in retrospect. A combination of ventricular inversion and normally related and connected great arteries is diagnostic of this malformation.

Summary of previously reported cases. Although 10 of the 14 patients reported on underwent cardiac catheterization, the correct anatomic diagnosis was made in only 1. The most commonly made but erroneous diagnoses included complete transposition of the great arteries (4), cor-

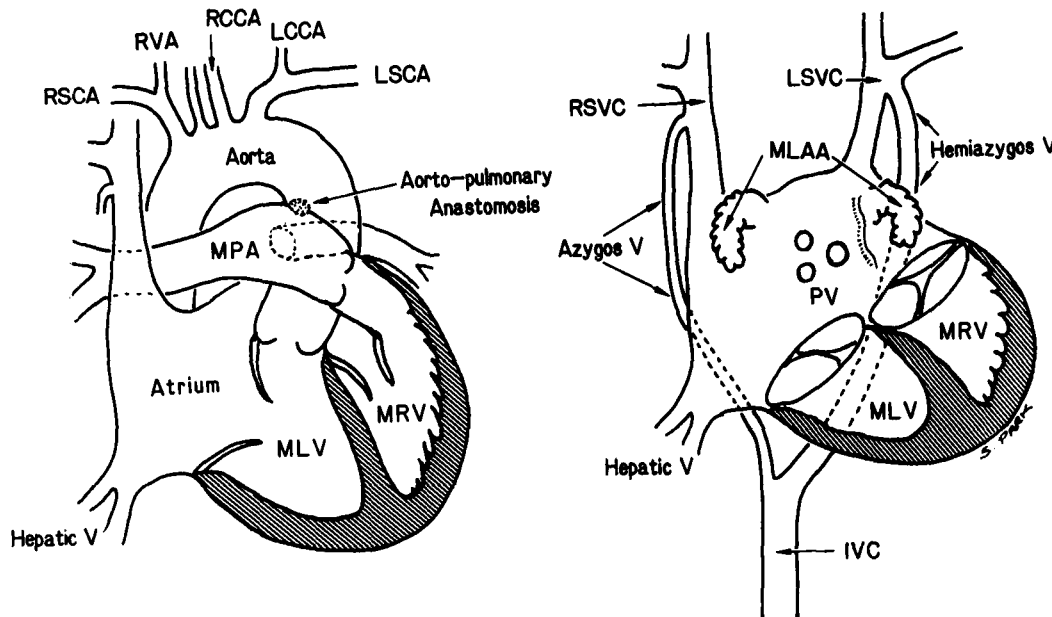


Figure 3. Diagrammatic illustration of cardiac anatomy based on angiographic and operative observations. IVC = inferior vena cava; LCCA = left common carotid artery; LSCA = left subclavian artery; LSVC = left superior vena cava; MLAA = morphologic left atrial appendages; MLV = morphologic left ventricle; MPA = main pulmonary artery; MRV = morphologic right ventricle; PV = pulmonary vein; RCCA = right common carotid artery; RSCA = right subclavian artery; RSVC = right superior vena cava; RVA = right vertebral artery; V = vein.

rected transposition of the great arteries, Taussig-Bing anomaly, tetralogy of Fallot and partial endocardial cushion defect. The clinical course of these patients was generally stormy. Six were profoundly cyanotic or in congestive heart failure, or both, and died without surgical intervention, all within the first 6 months of life. The remaining eight patients underwent 14 separate surgical procedures including atrial septectomy, pulmonary artery banding and various types of systemic-pulmonary artery anastomosis. One patient underwent Baffle's procedure unsuccessfully. Two of three patients, including our case, survived Mustard operation.

In the 14 reported cases there was a high incidence of an abnormal relation between visceral situs and cardiac position, levocardia with situs inversus in three isolated dextrocardia in two and mirror image dextrocardia in one. Anomalous systemic venous drainage was noted in only three patients, absence of the inferior vena cava with azygos continuation in three and persistent left superior vena cava draining into the left atrium in one. A high incidence of associated intracardiac anomalies was also noted. The ventricular septum was intact in four patients, while eight had a ventricular septal defect and two had a single ventricle. The atrial septum was intact in eight patients, three had a

common atrium and three a secundum atrial septal defect. The left-sided morphologic tricuspid valve was cleft or stenosed in seven patients, and the right-sided mitral valve was abnormal in three (stenosis, atresia and cleft, respectively). Hypoplasia of the left-sided morphologic right ventricle occurred in five patients, while the right-sided morphologic left ventricle was hypoplastic in one and absent in another (single right ventricle). A right-sided aortic arch was seen in five patients, four of whom had situs inversus. Abnormalities of other organs were rare and there were no associated chromosomal abnormalities or mental deficiencies in these patients.

Differentiation from complete transposition of the great arteries. This anomaly closely resembles complete transposition of the great arteries clinically and hemodynamically. In both situations, systemic venous blood enters the aorta and pulmonary venous blood enters the pulmonary artery. The hemodynamic alterations in transposition of the great arteries are a result of ventriculoarterial discordance. In our case atrioventricular (AV) discordance (isolated ventricular inversion) was the cause. In precise anatomic terms, our patient did not have AV discordance because there was left atrial isomerism and a common atrium. However, the orientation of the venous return to the AV valves resulted in physiologic AV discordance, which made a Mustard operation technically simple and quite effective in correcting the hemodynamic abnormality. Because the morphologic left ventricle was and postoperatively remains the systemic ventricle and the mitral valve the systemic AV valve, an excellent long-term result may be anticipated.

In conclusion, ventricular inversion with ventriculoarterial concordance is frequently associated with complex intracardiac lesions. Premortem diagnosis has been reported

in only one patient and successful surgical repair in only two. Two-dimensional echocardiography should lead to the correct diagnosis, but associated cardiac lesions should be carefully defined by cardiac catheterization and angiography.

Addendum

Since submission of this manuscript, the patient has undergone cardiac catheterization 12 months postoperatively. This study demonstrated an excellent hemodynamic result with normal pulmonary artery pressure (30/10 mm Hg, mean 18) and no ventricular outflow pressure gradients. There was no evidence of systemic or pulmonary venous obstruction.

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